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Research Article

A RARE CASE OF LEFT DIAPHRAGMATIC DOME AGENESIS IN AN ADULT

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ABSTRACT

In this paper we describe a case of a 31-yearold man having as antecedents of a subtotal colectomy at the age of 7 months for volvulus on common mesentery. He suffered from chest pain and dyspnea for 4 years. The paraclinical examinations posed the diagnosis of a left diaphragmatic hernia; it was during surgical exploration that we discovered that it was a complete agenesis of the left diaphragmatic dome.

INTRODUCTION

Diaphragm malformations are affections mainly of newborn, associated with other malformations and responsible for around 40% -50% mortality [1, 2]. Complete diaphragmatic dome agenesis (DA) is the largest diaphragmatic defect. The defect may be associated with herniation of the abdominal contents into the thoracic cavity and pulmonary hypoplasia, that results in progressive respiratory and cardiocirculatory failure and death of the neonate.

The DA is a rare pathology, and a late presentation of DA is exceedingly rare, her discovery is often fortuitous because it is often asymptomatic [3, 4]. There are only around twenty previous reports of adult DA in the literature [5, 6, 7]

In this study we describe the case of a young man patient suffering from chest pain and dyspnia for many years. The surgical technique, which included the use of a single layer ePTFE mesh to reduce the risk of either intrathoracic or intra-abdominal postoperative adhesions, is extensively described.

Case report

A 31-year-old man, complaining of chest pain in left hemithorax and dyspnea. At the age of 7 months he underwent a subtotal colectomy for volvulus on common mesentery. The patient reported no previous trauma or thoracic surgery, and her family history was normal.

A physical examination revealed a chest deformation, a decrease in the vesicular murmur in the left hemithorax,

Chest radiograph showed presence of hernia contents in thorax [Fig.1].

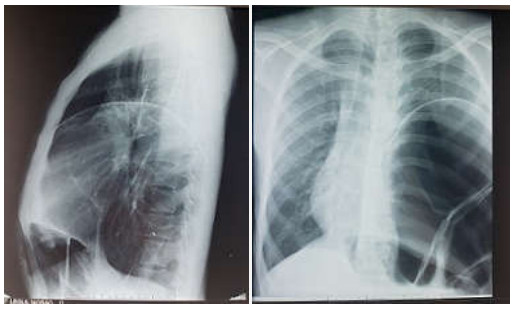

Fig 1 chest radiograph (postero-anterior view (R), left profil (L) showing the presence of hernial contents

CT scan showed the presence of intestinal gas and diminished pulmonary volume in the left thorax cavity (Fig. 2) and confirmed the herniation of the stomach, colon, small bowel and spleen. Spirometry and blood gas analysis were normal.

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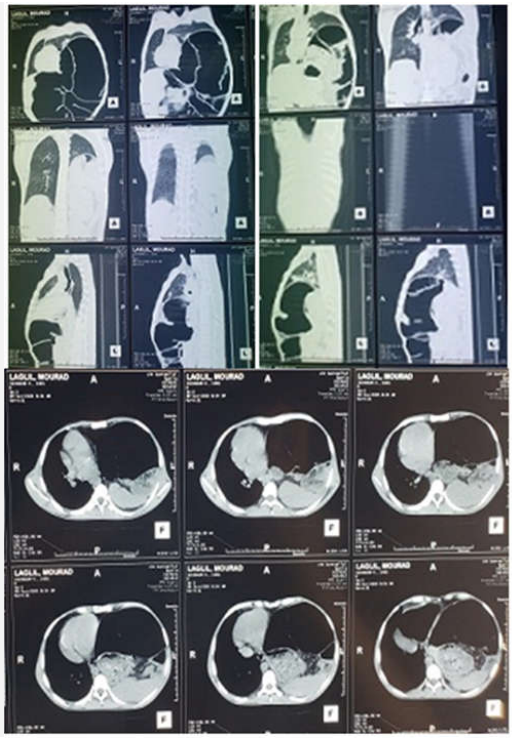

Fig 2 Chest CT scan (scout view): diminished pulmonary volume in the left hemithorax and herniation of stomach and colon. Chest CT scan (axial view): absence of left hemidiaphragm and herniation of stomach, small bowel, colon and spleen.

Surgical treatment was performed by a middle laparotomy straddling the umbilicus.

During abdominal exploration, we did not find any vestige of the left hemidiaphragm, except a thin antero-medial muscle border present rim, and the stomach, the spleen, part of the small bowel and the great omentum were dislocated into the left thorax (Fig 3) With left pulmonary hypoplasia.

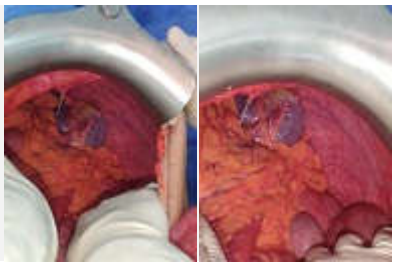

Fig 3 laparotomy view showing the diaphragm absence in all aspects: lateral, posterior, and anterior.

Once these viscera were replaced into the abdomen, the repair was performed with a 2-mm thick expanded polytetraXuoroethylene (ePTFE) soft tissue patch shaped upon the diaphragmatic defect. The patch was anchored without tension circumferentially to the very small diaphragm remnant, to the rib cage, to the rim of the endothoracic fascia and muscle by multiple running and a tapered point non-cutting needle (Fig. 4).

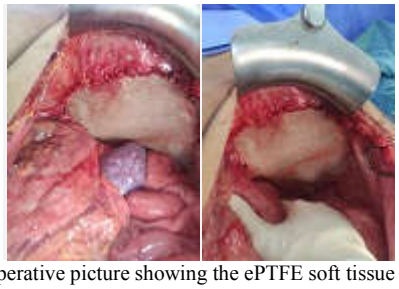

Fig 4 Intraoperative picture showing the ePTFE soft tissue patch in site

The patient received intravenous administration of analgesic drugs during the early postoperative 72 h. No early major and late postoperative complications were observed.

DISCUSSION

Diaphragm malformations are pathologies of the newborn, they are rare with an incidence of 0.8 - 5 / 10,000 births and a predominantly male [8, 9]. Among others congenital pathologies complete agenesis diaphragm and accessory diaphragm. Detected either by cardio-respiratory distress which can be fatal, or in an array malformative.

In early pregnancy, the embryonic pleuroperitoneal membrane and transverse septum join to begin diaphragm formation. In the third week of gestation, the association of the transverse septum with the dorsal mesentery of the foregut gives rise to two openings in which the thoracic and abdominal contents meet. In the ninth week of pregnancy, these openings close [10]. Thus, any process that prevents the closing of these channels can lead to defects in the diaphragm, including a congenital diaphragmatic hernia and DA [11].

Specifically, the pathogenesis of DA is still not clear. Some evidence suggests that it occurs due to improper development of the multiple embryonic sources of the diaphragm (the transverse septum, peritoneal folds and the somites [10].

In neonates, the clinical course of diaphragmatic dome agenesis is well documented; however, there is little significant information regarding this condition in adulthood. DA is extraordinarily rare in adults because pulmonary hypoplasia leads to gradual respiratory failure and, consequently, to the death of the individual after birth [12].

The adult patients are asymptomatic for a long time period without developing ventilatory insuficiency, perhaps and paradoxically just because of the associated lung hypoplasia; this should allow for more favorable ventilation-perfusion matching because it should minimize the large impairment of gas exchange that usually leads to death in neonates [6, 7, 13].

Chest X-ray or contrast upper gastrointestinal radiography, computed tomography (CT), and magnetic resonance imaging (MRI) are the imaging modalities used to establish a diagnosis of diaphragmatic dome agenesis. In cases where patients have symptoms of obstruction due to an intrathoracic visceral hernia, chest X-rays are used to aid diagnosis. A CT scan of the chest and abdomen is necessary in cases of solid organ herniation or negative X-ray findings. CT and three-dimensional reconstruction are the most advanced resources currently available; however, even CT cannot be conclusive, and in some cases, it may be necessary to use sagittal or coronal MRI [14].

Individuals suffering from congenital DA can remain asymptomatic for decades. When symptomatic, early surgical intervention may prove beneficial for the patient.

The issue of approach which should be adopted in adult patients with DA is controversial. Often, those exhibiting right diaphragmatic dome agenesis do not require surgery because the liver prevents the occurrence of a visceral hernia.

However, left DA may more often require reconstruction. Patients presenting symptoms such as dyspnea, recurrent pleural effusion, or organ herniation require a surgical approach, regardless of the location of the defect.

A thoracotomy in the fifth or sixth intercostal lateral space offers excellent access, although a transabdominal approach may be used if necessary [15].

The surgical treatment of diaphragmatic defects can be done by different techniques, but the repair must be performed with no stretching and minimal anatomical and functional alterations [15].

Among the different factors influencing the technical choice, the size of the defect is the most important [13, 16].

In case of patients with a large defect or agenesis of the hemidiaphragm, a direct suture may be impossible because of the absence of diaphragmatic tissue [15] or may lead to excessive tension. we can use prosthetic reconstructions.

An open question related to the management of diaphragmatic defects in adult patients is when and how to operate. As to the indication for surgery, in case of a small diaphragmatic defect there is substantial agreement toward an immediate repair even in asymptomatic subjects, because of the risk of bowel incarceration and strangulation [10, 12, 15].

Debate arises in case of asymptomatic adults patients with large diaphragmatic defects, because the bowel complications are rare thanks to the free movements of the herniated viscera through the large defect [14, 15].

CONCLUSION

Diaphragmatic malformations remain rare, including agenesis of the diaphragm which is exceptional in adults.

Despite the complexity and great diversity pathology of the diaphragm, this anomaly must be known because it is often well tolerated with non specific symptoms, sometimes remaining totally latent, imagery is essential but the etiology is difficult to specify by radiology alone.

The prognosis is generally good but depends on the associated malformations.

References

1. Journal de Radiologie- Organeofficiel de la Société Française de Radiologie(SFR) et du Collège des EnseignantsenRadiologie de France (CERF) Vol 78 – N° 4 – Avril 1997 p. 309-309. 2.
2. Muraskas JK, Husain A, Myers TF, Anderson CL, Blak PR. An association of pulmonary hypoplasia with unilateral agenesis of the diaphragm. J PediatrSurg 1993; 28:999-1002.
3. VALCKE J., DABADIE A.; P.GORINCOUR G. JFR – Paris – Octobre 2011.Service d'ImageriePédiatrique et Prénatale Centre Pluridisciplinaire de Diagnostic PrénatalHôpitald'Enfants de la Timone, Marseille, France. JFR – Paris – Octobre 2011.
4. VinodBhan, P. Rajagopal, Kiran Kumar and K.H. Raghavendra. Agenesis of the Hemidiaphragm: A Rare Presentation in an Adult. *Indian J Chest Dis Allied Sci* 2013; 55:109-111.
5. Singh G, Bose SM (1993) Agenesis of hemidiaphragm in adults. Aust NZ J Surg 63:327–328
6. Tzelepis GE, Ettensohn DB, Shapiro B, McCool FD (1988) Unilateral absence of the diaphragm in an asymptomatic adult. Chest 94:1301–1303
7. Anderson CD, Eliason J, Roberts JR, Sharp KW (2003) Bilateral diaphragmatic agenesis in an adult complicated by a paraduodenal hernia. Am Surg 69:523–525
8. Colvin J, Bower C, Dickinson JE, Sokol J. Outcomes of congenital diaphragmatic hernia: a population-based study in Western Australia. Pediatrics. 2005; 116(3):e356–363.
9. McGivern MR, Best KE, Rankin J, Wellesley D, Greenlees R, Addor MC, Arriola L, de Walle H, Barisic I, Beres J, *et al*. Epidemiology of congenital diaphragmatic hernia in Europe: a register-based study. Arch Dis Child Fetal Neonatal Ed. 2015; 100(2):F137–144.
10. Ali SA, Haseen MA, Beg MH. Agenesis of right diaphragm in the adults: a diagnostic dilemma. *Indian J Chest Dis Allied Sci*. 2013;56:121-123.
11. Jha A, Ahmad I, Naseem I. A rare case of partial diaphragmatic agenesis with thoracic liver herniation and anteriorly displaced intrathoracic kidney in an adult diagnosed by displaced diaphragmatic crus. Hernia. 2014;18:893-896.
12. Medina CB, Pêgo-Fernandes PM, Jatene FB. Pneumotóraxrecidivantesecundário à agenesia do diafragma. *J Pneumol*. 2002;28:48-50.
13. Anderson CD, Eliason J, Roberts JR, Sharp KW (2003) Bilateral diaphragmatic agenesis in an adult complicated by a paraduodenal hernia. Am Surg 69:523–525
14. Pousios D, Panagiotopoulos N, Argyriou P, Piyis A. Diagnosis and surgical management of diaphragmatic agenesis in an adult: report of a case. Surg Today. 2010;40:357-359.
15. Menezes SLS, Chagas PSC, Macedo-Neto AV, Santos VCT, Rocco PRM, Zin WA (2000) Suture or prosthetic reconstruction of experimental diaphragmatic defects. Chest 117:1443–1448
16. Atkinson JB, Poon MW (1992) ECMO and the management of congenital diaphragmatic hernia with large diaphragmatic defects requiring a prosthetic patch. *J Pediatr Surg* 27:754–756

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